<Case Report>Cerebellar Infarctions Secondary to Cranio-Cervical Anomalies : A Case Report

AUTHOR(S):

ANDO, SEIICHI; MATSUI, YUZURU; FUJII, JUNICHI; NAKAHARA, SHINNOUSUKE; MORITAKE, KOUZO

CITATION:

ANDO, SEIICHI ...[et al]. <Case Report>Cerebellar Infarctions Secondary to Cranio-Cervical Anomalies : A Case Report. 日本外科宝函 1994, 63(4): 148-154

ISSUE DATE:

1994-07-01

URL:

http://hdl.handle.net/2433/203635

Arch Jpn Chir 63(4), 148～154, Juli., 1994

症　　例

Cerebellar Infarctions Secondary to Cranio-Cervical Anomalies: A Case Report

Seiichi Ando*, Yuzuru Matsui**, Junichi Fujii, Shinnosuke Nakahara*** and Kouzo Moritake****

*Department of Neurosurgery
**Department of Orthopaedics, Unnan General Hospital, Shimane, Japan
***Department of Orthopaedics, Okayama University Medical School, Okayama, Japan
****Department of Neurosurgery, Shimane Medical University, Izumo, Japan

Received for Publication, March. 24, 1994

Abstract

We report a case of cerebellar infarctions which occurred in the territories of the bilateral posterior inferior cerebellar arteries. This case was complicated with cranio-cervical anomalies composed of assimilation of the atlas, atlanto-axial dislocation, and basilar impression.

The 40-year-old male patient had no detectable risk factors predisposing to atherosclerotic arterial occlusion or cardiogenic embolism, and there were no angiographic findings of atherosclerosis. It was, therefore, postulated that the cerebellar infarctions were secondary to those cranio-cervical anomalies. The developing mechanism is discussed.

Congenital anomalies at the cranio-cervical region rarely lead to the posterior fossa infarction [1-19]. We report a case of cerebellar infarctions which were thought to be secondary to multiple cranio-cervical anomalies.

Case Report

A 40-year-old man with no previous disease was suddenly attacked by dizziness, unsteady gait, and headache. Nausea and vomiting followed them immediately. The patient was admitted to our hospital on the next day when he became slightly drowsy and unable to walk.

The clinical contitions on admission were as follows. Pulse (68/min) and respiration (16/min) were regular. Blood pressure was 120/84 mm Hg. Body temperature was 36.4°C. Physical findings of the heart, lungs, and abdomen were normal. He was slightly drowsy (14 on the Glasgow

Key words: Vertebrobasilar infarction, Cerebellar infarction, Cranio-cervical anomaly, Assimilation of the atlas, Atlanto-axial dislocation, Baislar impression.
索引語: 椎骨脳底動脈領域脳梗塞，小脳梗塞，頭蓋頚椎移行部先天奇形，環椎癒合，環軸脱臼，頭蓋底嵌入症.
Present Address: Department of Neurosurgery, Unman General Hospital, 96-1 Daito-cho Iida, Ohara County, Shimane Prefecture, Japan (zip cord 699-12)

Coma Scale). Gross muscle strength of 4 extremities was normal, but he could not stand up. His speech was slurred. Spontaneouss horizontal nystagmus of both eyes was seen. The gag reflex was decreased bilaterally. Tendon reflexes of 4 extremities were symmetrically increased. The Babinski sign was positive bilaterally. Ataxia of 4 extremities and adiadochokinesia were demonstrated.

Electrocardiogram showed no pathological findings. Echocardiogram revealed no organic cardiac abnormalities.

The red cell count, Hb, and Ht were $460 \times 10^4/mm^3$, 13.3 g/dl and 39.8%, respectively. The white cell and platelet counts were $8,600/mm^3$ and $295 \times 10^3/mm^3$ Prothrombin time, active partial thrombin time, fibrinogen and antithrombin III were 12.5 sec (75.1%), 28.7 sec, 181 mg/dl and 86%, respectively. Fasting blood sugar was 89 mg/dl. Total cholesterol, HDL-cholesterol, and triglycerides were 147 mg/dl, 42 mg/dl, and 75 mg/dl, respectively.

CT on admission revealed that each cerebellar hemisphere separately had a lesion of ill-defined homogeneous low density. MRI showed that each cerebellar hemisphere individually had a large homogeneous intensity lesion, where the intensity was low on T1-weighted and high on T2-weighted images (Fig. 1). The cerebellar lesions on CT almost corresponded to those on MRI in size and site. Vertebral angiography (VAG) on the same day revealed following findings (Fig. 2). Bilateral vertebral arteries (VAs) were fully seen in their whole running routes, though the left one was hypoplastic. The left posterior inferior cerebellar artery (PICA) was not visible from its orifice and the right PICA was blocked at its posterior medullary segment (Fig. 2, arrow marks). The basilar, bilateral anterior inferior cerebellar, and bilateral superior cerebellar arteries were fully shown. All arteries mentioned above did not exhibit atherosclerotic appearances such as stenosis, dilatation, tortuosity, or intimal irregularity. It was confirmed by CT, MRI, and angiography that the vast infarctions took place in the bilateral PICA territories.

Assimilation of the atlas[9,20] was suspected on the lateral view of a skull X-ray, and tomography

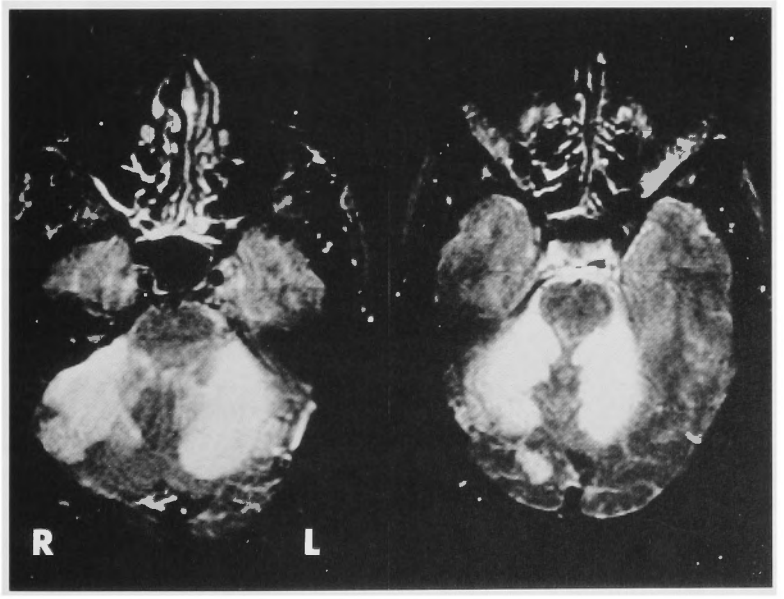

Fig. 1 T2-weighted MRI showed a large homogeneous high-intensity area in each cerebellar hemisphere.

demonstrated its existence (Fig. 3A). Atlanto-axial dislocation was revealed on the lateral view of the spinal X-ray films, when the head was kept anteriorly-flexioned or neutral (Fig. 3B)[9,20]. Basilar impression was demonstrated, when the head was kept posteriorly-flexioned (Fig. 3B)[9,20]. Chiari malformation and syringomyelia, which are occasionally associated with these cranio-cervical anomalies[9,20], were not seen on the sagittal views of MRI.

We speculated that the infarctions in the bilateral PICA territories were induced by the triplicated cranio-cervical anomalies. In order to prevent recurrence of the posterior fossa infarction, we performed, according to the method established by MAGERL and a colleague, trans-articu-lar screw fixations of the bilateral atlanto-articu-lar joints[21,22]. Fusion between the occipital bone and the lamina of the C2 was simultaneously done with an iliac bone graft. Postoperative X-ray films revealed that the pathological conditions due to atlanto-axial dislocation and basilar impression were completely corrected. Fusion was successfully accomplished.

The patient regained the ability to walk 3 months after the operation.

Discussion

It has been established that there is a causative relationship between cranio-cervical anomalies and infarction of the vertebrobasilar system[1-19].

The atlanto-axial joint moves excessively in the patients with atlanto-axial dislocation[3,9]. The cranio-cervical joint extremely increases in mobility in the patients with atlanto-axial dislocation or basilar impression[1,19,20]. Pathological hypermobility of the cranio-cervical and/or atlanto-axial joints could expose the uni- or bilateral VAs, which run tortuously through these two joints, to me-

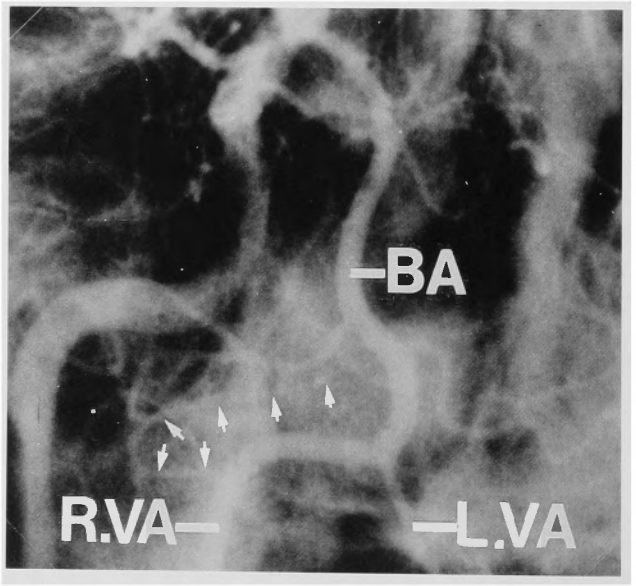

Fig. 2 **Right transbrachial retrograde VAG.**
Abbreviations: R.VA; the right vertebral artery, L.VA; the left vertebral artery, BA; the basilar artery.
Arrow marks: the right posterior inferior cerebellar artery.

chanical stress such as compression, stretching, angulation or kinking (Fig. 4)[1,3,5,6,9,10,14,16,18,19]. The mechanical stress is thought to be responsible for initiating the infarction in the vertebro-basilar system (Fig. 4)[1,6,14,18]. Organically, it induces intimal injuries, intimal oedema, or vasoconstriction [1,3-5,9,10,11-17]. Those kinds of mechanical stress mentioned above might directly lead to VA obliteration, which could result in the infarction (Fig. 4). Progression of local thrombosis at the intima-injured part would obstruct VA in situ (Fig. 4)[1,10,12,13,18]. Some authors have suggested that the emboli formed at the injured intima would obstruct the arteries distal to VA (Fig. 4)[3,10,11,14-17]. Other authors have reported that embolism occurred secondary to emboli from a pseudoaneurysm that developed at the intima-injured area (Fig. 4)[4,5,13,17] Functionally, on the other side, the mechanical stress causes a decrease or stasis in the blood flow of VA[1,6-9,18,19]. Intermittent vertebro-basilar insufficiency in a form of transient ischaemic attack is surmised to occur in this situation (Fig. 4)[1,8,9,18,19]. Furthermore, it is also believed that a decrease or stasis in the blood flow of VA could make small emboli and eventually accomplish the completed stroke (Fig. 4)[6-8]. It is said that, the causing mechanism being organic or functional (Fig. 4), the infarction is more likely to occur when the blood flow of the contralateral VA is, as was seen in our case, insufficient because of hypo- or aplasia[1,8,10,12,15]. In addition, it is suggested that assimilation of the atlas, which was demonstrated in our case, could bring about atlanto-axial dislocation or basilar impression[9,20].

The major cause of posterior fossa infarction in adult is atherosclerotic occlusion[3,15,24]. Another important one is embolism from the heart or from the occluded extracranial artery[3,15,24]. It seems unlikely that atherosclerotic occlusion or embolism took place in our case, since there were no detectable risk factors predisposing to atherosclerosis or embolism. Furthermore, no angiographic appearances of atherosclerosis were observed. We speculate that, in our case, hypermobility of the atlanto-axial joint participated in the initiation of infarctions in the territories of bilateral PICAs. The infarction can be supposed to have taken one of the hypothesized courses in Fig. 4. The infarctions secondary to direct VA obliteration, local thrombosis of VA, and embolism based on pseudoaneurysm were denied on VAG. However, we could not elucidate the developing mechanism in greater detail.

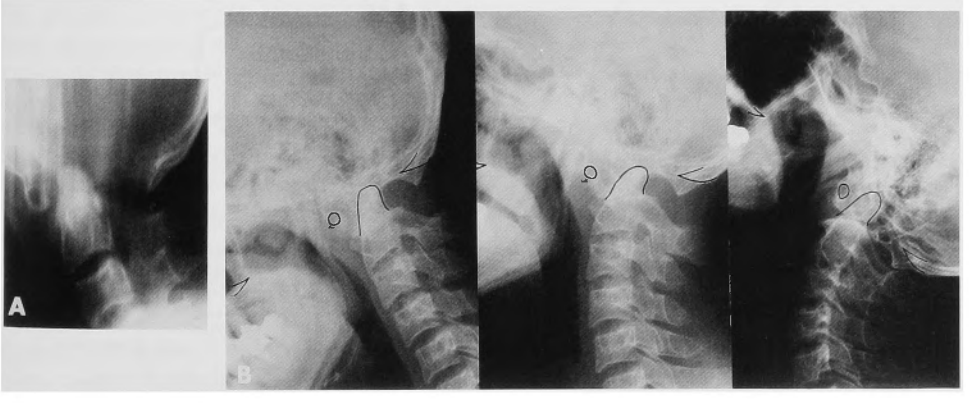

Fig. 3 A: Tomography of the cranio-cervical junction. Assimilation of the atlas was demonstrated. B: The lateral views of the spine X-rays. The distance between the posterior aspect of the anterior arch of the atlas and the anterior surface of the odontoid process was 7 mm in the anteriorly-flexioned position and 8 mm in the neutral position. An X-ray film taken in the posteriorly-flexioned position demonstrated that the tip of the odontoid process was situated 6 mm above Chamberlain's line and 7 mm above MacGregor's line.

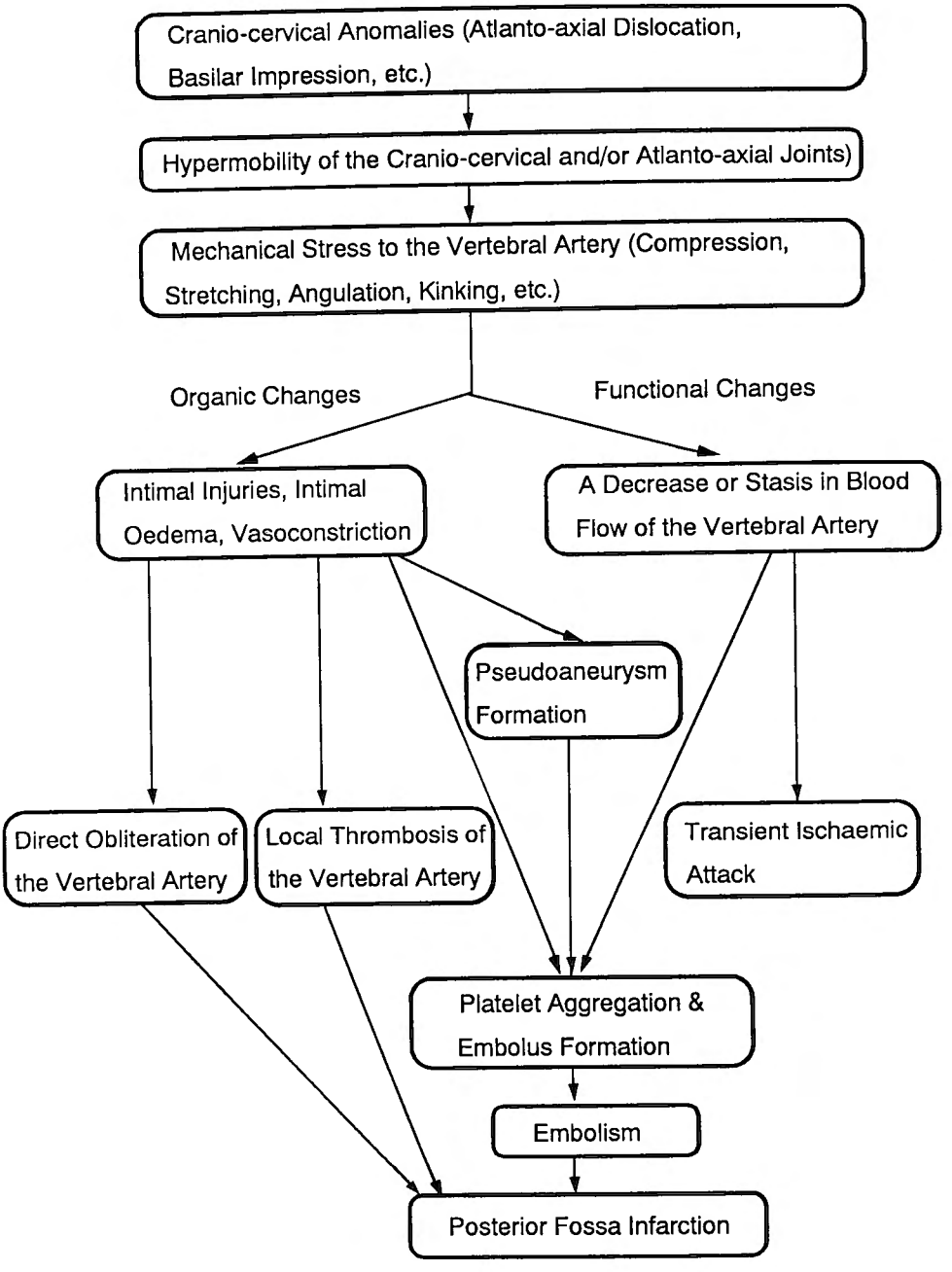

Fig. 4 Schematic Illustration of Posterior Fossa Infarction Due to Cranio-cervical Anomalies.

The posterior fossa infarction due to cranio-cervical anomalies usually develops in middle or later life[9,25]. Generalized arteriosclerosis and arachnoidal thickening at the cranio-cervical and atlanto-axial joints, both of which physiologically begin in middle age and increase in severity with age, are thought to be related to middle-aged or later onset[9,25]. Namely, generalized arteriosclerosis

takes part in decompensation of the vertebrobasilar arterial system for decreased blood flow. Arachnoidal thickening is believed to exaggerate the mechanical stress to VA.

Surgical fixation of the cranio-cervical or atlanto-axial joint is the only way to prevent recurrence of vertebro-basilar infarctions secondary to cranio-cervical anomalies[1,4,6,7,14,17,18]. We used a modification of MAGERL's method, which is advantageous for fixing the atlanto-axial joint three-dimensionally[21,22], and achieved a satisfactory results.

References

1) Bell HS: Basilar artery insufficiency due to atlanto-axial instability. *Am Surg* **35**: 695–700, 1969.
2) Bernini FP, Elefante R, Smaltino F, Tedeschi G: Angiographic study on the vertebral artery in cases of deformities of the occipito-cervical joint. *Am J Roent Radiat Ther Nucl Med* **107**: 526–529, 1969.
3) Caplan LR, Zarins CK, Hemmati M: Spontaneouss dissection of the extracranial vertebral arteries. *Stroke* **16**: 1030–1038, 1985.
4) Chakera TMH, Anderson JEM, Edis RH: Case reports Atlanto-axial dislocation and vertebral artery aneurysm. *Brit J Radiol* **55**: 863–864, 1982.
5) Davidson KC, Welford EC, Dixon GD: Traumatic vertebral pseudoaneurysm following chiropractic manipulation. *Radiology* **115**: 651–652, 1975.
6) Ford FR: Syncope, vertigo and disturbances of vision resulting from intermittent obstruction of the vertebral arteries due to defect in odontoid process and excessive mobility of the second cervical vertebra. *Bull Hopkins Hosp* **91**: 168–173, 1952.
7) Fraser RAR, Zimbler SM: Hindbrain stroke in children caused by extracranial vertebral artery trauma. *Stroke* **6**: 153–159, 1975.
8) Grossmann RI, Davis KR: Positional occlusion of the vertebral artery: A rare cause of embolic stroke. *Neuroradiology* **23**: 227–230, 1982.
9) Hensinger RN: Congenital anomalies of the cervical spine, In: Rothman RH, Simeone FA, eds. *The Spine*, Vol 1, Philadelphia, W. B. Saunders Company, 1992, ed 3, 261–348.
10) Hope EE, Bodensteiner, Barnes P: Cerebral infarction related to neck position in an adolescent. *Pediatrics* **72**: 335–337, 1983.
11) Latchaw RE, Seeger JF, Gabrielsen TO: Vertebrobasilar arterial occlusions in children. *Neuroradiology* **8**: 141–147, 1974.
12) Marks RL, Freed MM: Nonpenetrating injuries of the neck and cerebrovascular accident. *Arch Neurol* **28**: 412–424, 1973.
13) Mas J-L, Boursser M-G, Hasboun D, Laplane D: Extracranial vertebral artery dissections: A review of 13 cases. *Stroke* **18**: 1037–1047, 1987.
14) Phillips PC, Lorentsen KJ, Shropshire LC, Ahn HS: Congenital odontoid aplasia and posterior circulation stroke in children. *Ann Neurol* **23**: 410–413, 1988.
15) Rosman NP, Wu JK, Caplan LR: Cerebellar infarction in the young. *Stroke* 23: 763–766, 1992.
16) Ross CA, Curnes JT, Greenwood RS: Recurrent vertebrobasilar embolism in an infant with Klippel-Feil anomaly. *Pediatr Neurol* **3**: 181–183, 1987.
17) Schneider RC, Crosby EC: Vascular insufficiency of brain stem and spinal cord in spinal trauma. *Neurology* **9**: 643–656, 1959.
18) Singer WD, Haller JS, Wolpert SM: Occlusive vertebrobasilar artery disease associated with cervical spine anomaly. *Am J Dis Child* **129**: 492–495, 1975.
19) Taylor AR, Chakravorty: Clinical syndromes associated with basilar impression. *Arch Neurol* **10**: 475–484, 1964.
20) Findlay GFG: Common malformations of the nervous system. In: Miler JD, ed. *Northfield's Surgery of the Central Nervous System*. Edinburgh, Blackwell scientific publications, 1987, ed 2, 574–596.
21) Grob D, Magerl F: Operative Stabilisierung bei Frakturen von C1 und C2. *Orthopäde* **16**: 46–54, 1975.
22) Hanson PB, Montesano P, Sharkey NA, Rauschning W: Anatomic and biochemical assessment of transarticu-lar screw fixation for atlantoaxial instability. *Spine* **16**: 1141–1145, 1991.
23) Taveras JM, Wood EH: Cerebral angiography. In: *Diagnostic Neuroradiology*, Vol 2, Baltimore, The Williams & Wilkins Company, 1976, ed 2, 543–986.

24) Kasa CS, Norrving B, Levine SR, Babikian VL, Chodosh EH, Wolf PA, Welch KMA. Cerebellar infarction. Clinical and anatomical observations in 66 cases. *Stroke* **24**: 76–83, 1993.
25) Caetano de Barros M, Farias W, Ataíde, Lins S: Basilar impression and Arnold-Chiari malformation. A study of 66 cases. *J Neurol Neurosurg Psychiat* **31**: 596–605, 1968.

和文抄録

頭蓋頸椎移行部の先天奇形により生じた小脳梗塞の1例

公立雲南総合病院 脳神経外科
安東 誠一

公立雲南総合病院 整形外科
松井 譲，藤井 淳一

岡山大学医学部 整形外科
中原進之介

島根医科大学 脳神経外科
森竹 浩三

両側の後下小脳動脈領域におこった小脳梗塞の1例（40歳，男性）を報告する．本例には環椎癒合，環軸脱臼，さらに頭蓋底嵌入症という3つの頭蓋頸椎移行部先天奇形が合併していた．

動脈硬化性動脈閉塞や心原性塞栓などをおこしうる危険因子は検索しえなかった．このため，本例の小脳梗塞は，頭蓋頸椎移行部の先天奇形を基盤として生じたものと思われた．発生機序につき詳細に検討した．